Acute Chest Syndrome in the Sickle Cell Patient

Children's Mercy KANSAS CITY | Evidence Based Practice

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Patient with Sickle Cell Disease experiencing Acute Chest Syndrome in the ED

Obtain history of:

- Prior Acute Chest Syndrome
- Asthma
- Fever
- Current chest pain
- Restrictive lung disease
- Nocturnal hypoxia
- Respiratory symptoms (cough, shortness of breath, dyspnea)
- Recent history of sedation without prior transfusion

If patient is known to Children's Mercy: Review Critical Information note

Initial pt. work-up to include:

- CBC w/differential, reticulocyte count, HbS level
- BMP, liver function panel, LDH
- Consider blood gas
- Blood culture if febrile, hypotensive or toxic-appearing
- Consider RVP and COVID-19 testing
- Type and Screen
- Chest X-Ray (2 views) for any respiratory symptoms, even in absence of hypoxia or abnormal lung findings on exam

Management of patient while in ED and transferred to either Hem/Onc resident service or PICU (dependent on patient status):

- Consult Hem/Onc
- Oxygenation:
  - Supplemental oxygen only if hypoxic (O_2 saturation: < 94% or > 4% below baseline if known chronic hypoxia)
  - Incentive spirometry every 2 hours with Respiratory Therapy while awake using age appropriate respiratory therapy (pinwheel and bubbles); encourage patient to accomplish hourly
  - Intermittent positive pressure breathing every 4 hours as indicated
  - Consider trial of bronchodilators (atrovent/albuterol) in patients with history of asthma or wheezing on exam. Follow asthma action plan in patients with history of asthma
- Encourage ambulation and activity
- Pain Management:
  - PO tylenol and NSAIDS
  - IV narcotic, refer to critical note for preferred narcotic
- Fluid Management:
  - Make NPO
  - Avoid fluid bolus as this may exacerbate pulmonary edema
  - Start IV fluids at three quarters to full maintenance rate
- Empiric antibiotics (Ceftriaxone and Azithromycin)
  - If patient allergic to cephalosporin, then consider Clindamycin
- Simple transfusion if Hgb > 2 gm/dL below baseline
  - Transfuse packed red blood cells (discuss volume with Hem/Onc consultant)
  - Sickle negative and cross-matched for C, E, Kell antigens*
  - Goal: Hgb 10 - 11 gm/dL
  - Labs after transfusion: CBC, retic count, and HbS

Is the patient exhibiting signs necessitating PICU care?

Yes →

- Transfer/Continue care in PICU
- Consult Hem/Onc Service
- Prepare for exchange transfusion

No ↓

Transfer to Hem/Onc Service

Management per Hem/Onc and Discharge Home

← Yes

Is patient stable for transfer to floor?

No →

References used to establish this care standard

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